Malignant tumors of nose and paranasal sinuses

By

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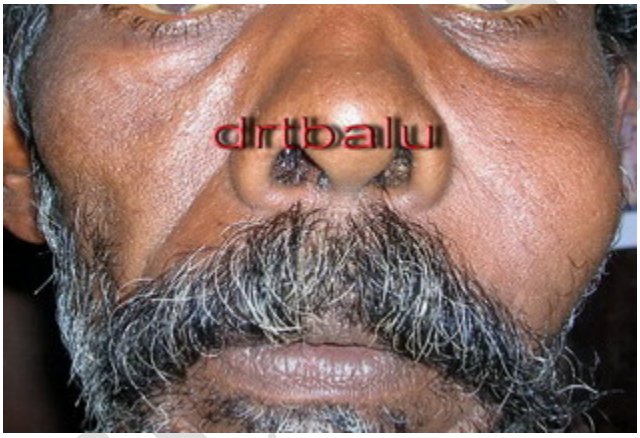

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Introduction:

Nose and paranasal sinuses represent two of the rarest sites of origin of head and neck cancer. They account for about 0.8% of all carcinomas. These tumors afflicting nose and paranasal sinuses could belong to a variety of histological types. Clinically these tumors are notorious for their late presentation and nodal metastasis. A strong index of suspicion is necessary to identify these lesions at an early stage. The proximity of these structures to orbit and skull base makes management of tumors in these areas that bit difficult.

Surgical anatomy:

1. The maxillary sinus is a box like structure bounded by orbit, nose, mouth, cheek pterygoid space and nasopharynx. Secretions from maxillary sinus may drain into the anterior ethmoid sinuses, and nose. Roots of teeth may be exposed to secretions within maxillary sinus if the floor of the sinus is dehiscent.

2. Lymphatic drainage of maxillary antrum is not clearly understood. Possibilities include:

a. Scanty posterior drainage into the retropharyngeal group of nodes

b. Lymphatics may directly drain to the jugulodigastric nodes

c. Anterior portion of the nasal cavity drains into submandibular group of nodes

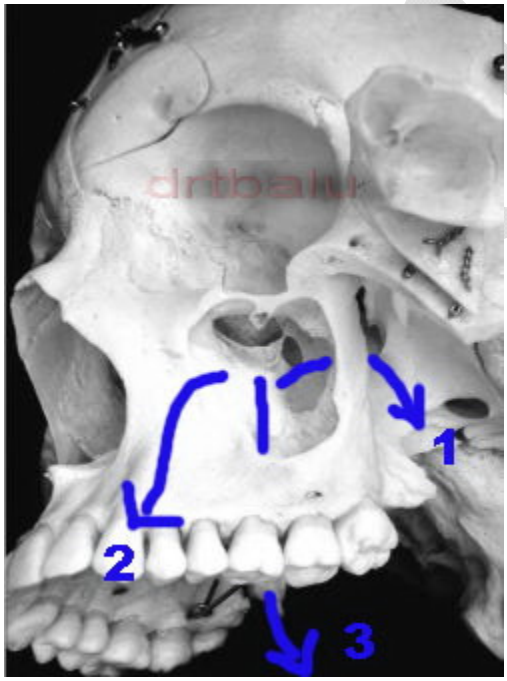


Figure showing lymphatic drainage of maxillary sinus: 1 - to retropharyngeal node

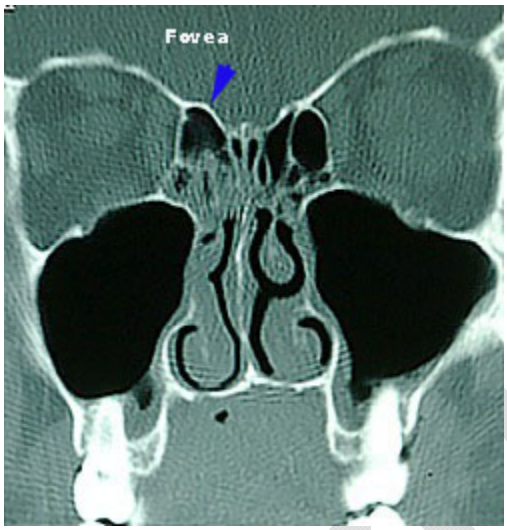
2 - Submandibular node 3 - Jugulodigastric node

Enlarged neck node seen in a patient with carcinoma involving the maxillary sinuses indicates the tumor has breached the maxillary antrum and has involved cheek area which is very rich in lymphatics.

3. The level of cribriform plate can be indirectly assessed by using naso frontal suture line as a surface marking. The cribriform plate is supposed to lie about 2.5 cm posterior to this suture line, separated by the upward extension of the anterior ethmoidal cells.

4. Posterior ethmoid cells lie just 3-5 mm away from the optic nerve canal.

5. The cribriform plate is bounded laterally by very thin roofed superior extension of ethmoidal cells i.e. known as fovea ethmoidalis



Coronal CT showing Fovea ethmoidalis

6. Anterior to the crista galli is a foramen for an emissary vein that comes from the superior saggital sinus.

7. Olfactory nerves ensheathed in dura pass through the perforations in the cribriform plate of ethmoid. The dura in this area is so tightly adherent to the bone it is virtually impossible to elevate and expose the roof of ethmoid without tearing the dura.

8. About 2.5 cms behind the posterior extent of cribriform plate lie the optic chiasma. Optic nerves should hence be visualized during cranifacial surgeries. Ethmoidal tumors extend to the brain through the dural sheath of optic nerves.

9. It should be borne in mind that the average distance between the columella and the optic nerve is 8.3 cms and the distance between columella and internal carotid artery is 8.6 cms.

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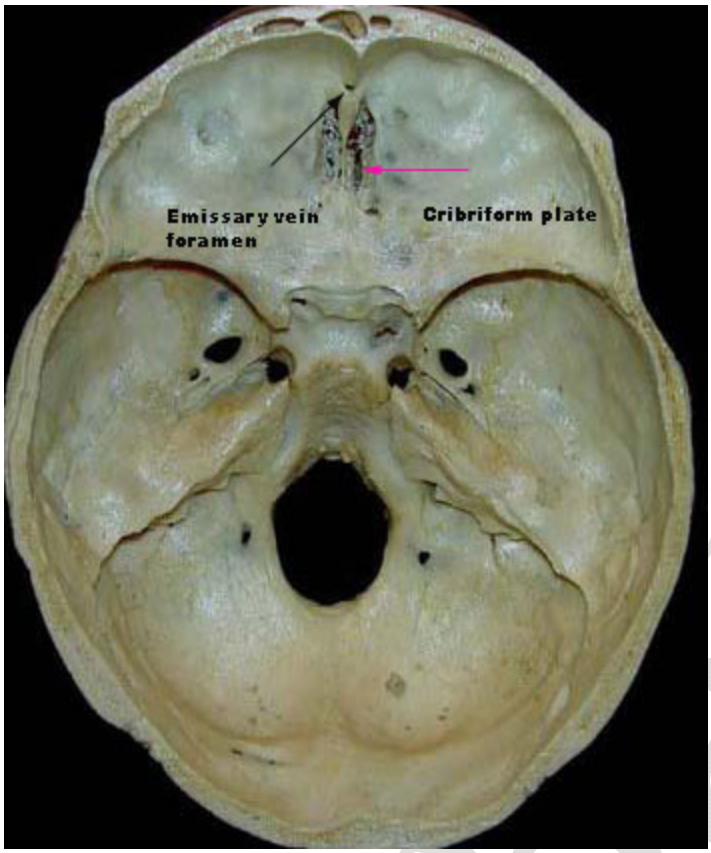

Figure of skull base

10. Ethmoidal sinus labyrinth is composed of 3 - 17 air cells. These cells extend posteriorly to become contiguous with the sphenoid sinus.

11. It should be borne in mind that the roof of the ethmoid is superior to the level of cribriform plate in the anterior cranial fossa.

12. Ethmoidal air cells are penetrated by the anterior and posterior ethmoidal arteries

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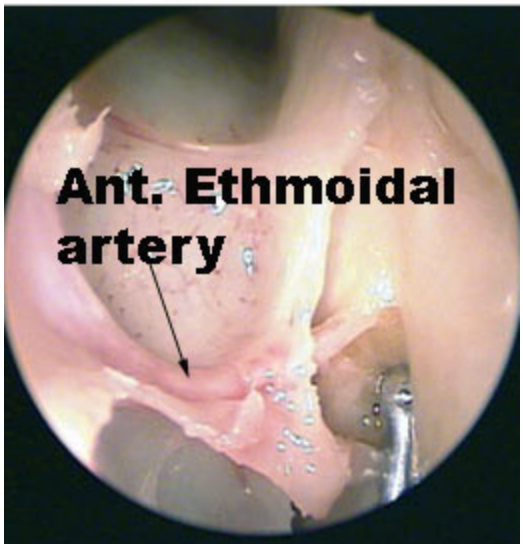

Endoscopic view of anterior ethmoidal artery

13. The anterior ethmoidal artery enters the ethmoidal sinus 4 cms behind the medial orbital ligament, and the posterior ethmoidal artery enters just 2 cms behind this. Posterior ethmoidal artery lies perilously close to the optic nerve.

14. During surgical removal of ethmoid maxillary complex - the three nasal turbinates are taken out, whereas lateral rhinotomy involves removal of middle and inferior turbinates.

15. Orbit is an important area which should be taken into consideration while surgically managing ethmo maxillary tumors. The eye is held in position by medial and lateral orbital ligaments. The ocular muscles superior oblique and inferior rectus are attached to the orbital periosteum. During surgery in this area it is of paramount importance to stay under the periosteum, inorder not to disturb these intraocular muscles.

16. Lacrimal sac - lies between the lacrimal bone and the frontal process of maxilla, just behind the medial orbital ligament. Lacrimal sac is usually damaged during maxillectomy. The presence of a resulting defect into the nasal cavity acts as a large dacryocystorhinostomy and the patients are usually asymptomatic.

17. The orbital periosteum is highly resistant to carcinomatous spread. In craniofacial resections the major advantage is that this orbital periosteum can be dealt with more accurately from above than below.

18. The superior and inferior orbital fissures offer ready made routes for entry and exit of tumors to and from the orbit.

19. The floor of the nasal cavity is formed by the hard palate; and the floor of the maxillary antrum is formed by the alveolus. The premolar and molar teeth are in intimate relationship to the floor of the maxillary sinus. To the posterior wall of maxillary sinus is

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attached the pterygoid plates. The pterygomaxillary fissure is the space between the pterygoid plates and maxillary sinus. The maxillary artery traverses through this space.

20. Infratemporal fossa - is the space behind the maxillary antrum. It is bound medially by the pterygoid plates, laterally by the zygomatic arch, coronoid process and ascending ramus of the mandible. Posteroinferiroly it connects to the parapharyngeal space, and superior to this fossa lie the sphenoid bone, foramen ovale and foramen spinosum. This fossa contains the mandibular division of the Vth cranial nerve, pterygoid muscles and maxillary artery along with venous plexus. Tumors involving this area could be primary or metastasis or spread from contiguous structures.

21. Lymphatic drainage: The lymphatics of the respiratory (non vestibular and olfactory) portion terminates in to the anterior jugular chain and the deeper nodes of the internal jugular vein. Regarding the lymphatic drainage of paranasal sinuses only that of ethmoids and maxillary sinuses are known with some degree of certainty. Lymphatics from these two sinuses join the lymphatics draining the lateral nasal wall and then continues to the retropharyngeal lymph nodes (first echelon). Lymphatics from the lateral wall of maxillary sinus drain into the submandibular and upper deep cervical chain. There is virtually no lymphatic communication between the antrum and buccal cavity, orbit. Any involvement of these areas could only be by direct extension.

22. Invasion of oral cavity by antral carcinoma could increase the risk of regional nodal metastasis.

Risk factors for sinonasal cancer:

Hard wood dust - adenocarcinoma

Soft wood dust - squamous carcinoma

Nickel refining - Chromium workers

Boot - shoe - textile workers

Isopropyl oil; volatile hydrocarbons

Snuff

Human papilloma virus

Sex preponderance - Male:Female = 2:1

Mean age of occurrence - 55 years

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Site of origin of cancer:

Almost 50% of all sinonasal malignant tumors arise from the lateral wall of the nose.

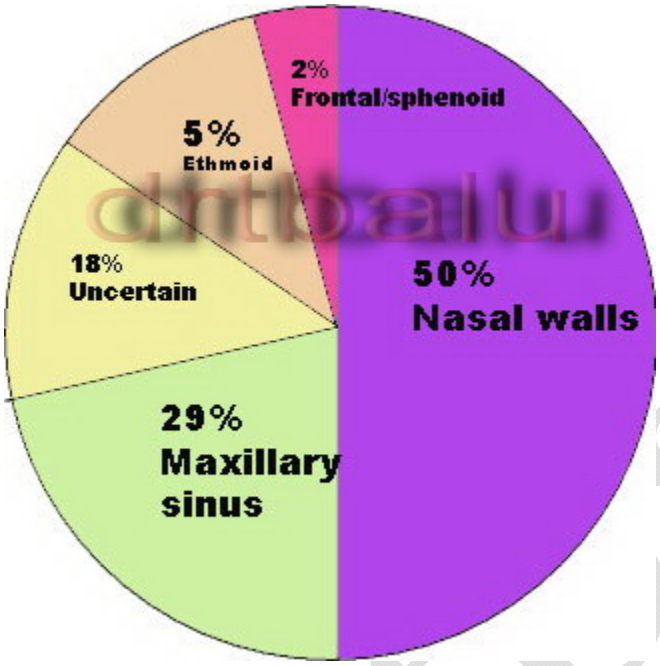

Figure showing the sites of occurrence of malignancy in nose and PNS

Classification of malignant tumors of nose and paranasal sinuses:

Majority of malignant tumors of nose and paranasal sinuses is squamous cell carcinoma.

a. Basal cell carcinoma

b. Squamous cell carcinoma

c. Tumors of minor salivary glands

d. Esthesioneuroblastoma

e. Lymphoreticular tumors

f. Plasmacytoma

g. Adenocarcinoma

h. Undifferentiated carcinoma

i. Malignant fibrous dysplasia

j. Malignant neurogenic tumors

k. Sarcoma

Ohngren's classification:

Ohngren in 1933 classified sinus tumors by drawing a line from the medial canthus of the eye to the angle of the jaw. This line was aptly named as "Ohngren's line". Structures postero superior to this line constitute the suprastructure while structures anteroinferior to this line constitute the infrastructure. He suggested that tumors located posterosuperior to this line has worse prognosis when compared to tumors located anteroinferior to this line.

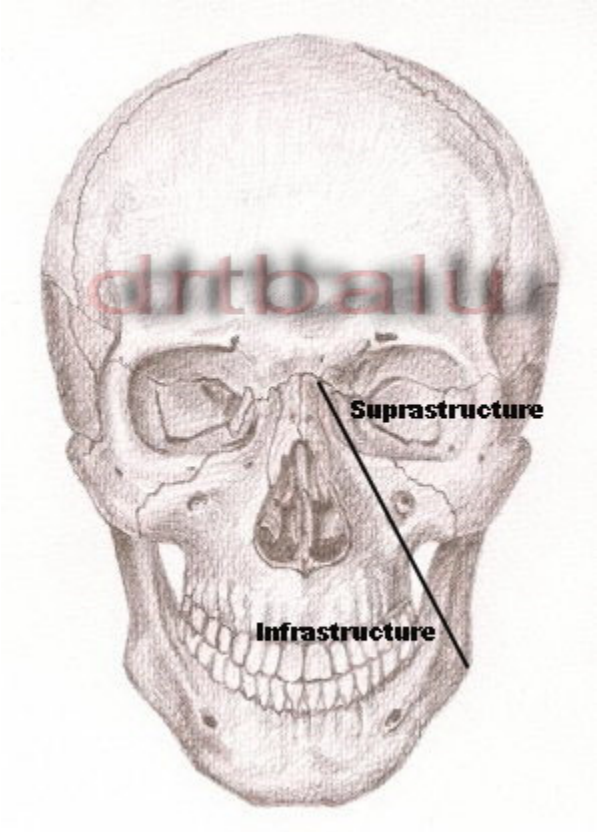

Figure showing Ohngren's line

Ledermann's classification:

Ledermann modified Ohngren's classification by using horizontal lines drawn through the floors of the orbit and antrum together with vertical lines extending down from the medial walls of the orbit to the floor of nasal cavity. This classification has the advantage of including tumors of the ethmoid sinus, nasal cavity as well as the maxillary antrum and hence comprehensive.

Suprastructure: contains ethmoid, sphenoid and frontal sinuses. This area also included the olfactory portion of the nasal cavity

Mesostructure: contains maxillary sinus and respiratory portion of the nasal cavity

Infrastructure: contains the alveolar process of maxilla

According to Ledermann malignant tumors of infrastructure carries the best prognosis, while tumors of meso structure carries intermediate prognosis while tumors of supra structure carries the worst prognosis.

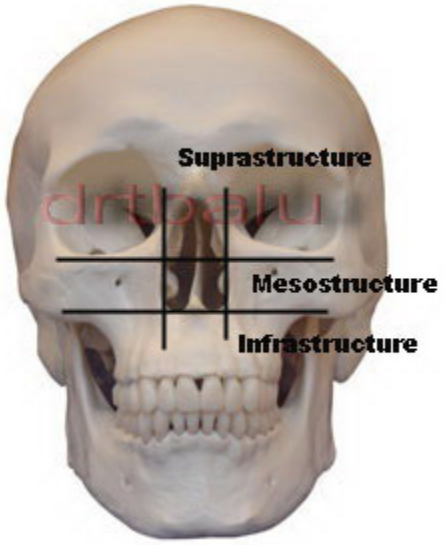

Figure showing Ledermann's classification

Harrison's classification of maxillary sinus tumors:

T1 - Tumor limited to the antral mucosa without evidence of bone erosion

T2 - Bone erosion without involvement of facial skin, orbit, ethmoids, or pterygopalatine fossa

T3 - Involvement of orbit, ethmoids, or facial skin

T4 - Tumor extension into nasopharynx, sphenoid sinus, cribriform plate, or pterygopalatine fossa

Types of malignant tumors involving the nose:

Basal cell carcinoma: Nose is the most common site for basal cell carcinoma of head and neck. Infact it is 30 times more common than squamous cell carcinoma. It commonly occurs during the 6th - 8th decades. Incidence of this tumor is almost equal in both sexes.

These tumors could either be circumscribed or infiltrative. If the lesion is ulcerative in nature, its borders could be either flat or rolled out. Its infiltration could be easily underestimated. Distant metastasis is rare. Wide excision of the tumor could be beneficial.

Squamous cell carcinoma: This is the second commonest malignant tumor involving the nose and paranasal sinuses. The fact that the lateral wall of the nose, maxillary sinus and ethmoidal sinuses are contiguous it is impossible to predict the origin of the carcinoma with certainty.

About 50% of these tumors arise from the maxillary antrum.

Squamous cell carcinoma involving the nasal vestibule commonly occurs in elderly men. This tumor is highly localized without any propensity to metastasis. This tumor can be managed with aggresive surgical excision / radiotherapy. Incontrast squamous cell carcinomas involving the columella of nose is really aggresive, with very high incidence of recurrence.

Tumor classification "Recent":

"T" classification

T - Primary tumor

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Maxillary sinus:

T1 - Tumor limited to antral mucosa with no evidence of bone destruction

T2 - Tumor with evidence of bone destruction, except for the posterior wall of maxilla, including extension into hard palate and / or middle meatus of nose

T3 - Tumor invading any of the following structures - Bone of posterior wall of maxillary sinus, subcutaneous tissues, skin of cheek, floor of medial wall of orbit

T4 - Tumor invading the orbital contents / or any of the following - cribriform plate, base of skull, nasopharynx sphenoid sinus and frontal sinus

Ethmoid sinus:

T1 - Tumor confined to ethmoid with or without bone destruction

T2 - Tumor extending into the nasal cavity

T3 - Tumor extends to anterior orbit and / or maxillary sinus

T4 - Tumor with intracranial extension, orbital extension including its apex, involving the sphenoid and / or frontal sinus / or skin of nose

Among the paranasal sinuses maxillary sinus is the one commonly affected by malignant tumors. About 80% of all paranasal cancers arise from the maxillary sinus. The ethmoidal sinuses come next. Primary carcinomas involving the frontal and sphenoid sinuses are rather rare.

Signs and symptoms of Maxillary sinus cancers:

Oral :

1. Unexplained pain in the maxillary teeth

2. Trismus

3. Fullness / expansion of the palate or alveolar ridge

4. Erosion through the wall of the sinus with protrusion into the oral cavity

5. Loose teeth / ill fitting dentures

6. Persistent oroantral fistula

7. Numbness over hard palate (greater palatine nerve)

8. Evidence of 9th & 10th cranial nerve palsies

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9. Palpable mass in the hard palate, buccal sulcus, or anterior antrum

Nasal:

1. Foul smelling nasal discharge

2. Stuffiness

3. Congestion

4. Epistaxis

5. Mass in the nasal cavity

Ocular:

1.Unilateral tearing

2. Diplopia

3. Fullness of lids

4. Orbital pain

5. Exophthalmos (common in ethmoidal tumors)

Facial:

1. Parasthesia over the infraorbital nerve

2. Swelling and pain over cheek

3. Facial asymmetry

In advanced tumors all these features may be seen.

Tumor spread:

Antral tumors spread to:

1. Nasal cavity

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2. Ethmoid

3. Orbit via the infra orbital fissure

4. Soft tissues of the cheek by eroding the anterior wall

5. Palate / alveolar ridge via the dental foramina

6. Buccal sulcus

7. Infratemporal fossa

Ethmoid tumors spread to:

1. Maxillary antrum

2. Orbit

3. Nose

4. Sphenoid (direct spread)

5. Anterior cranial fossa through fovea or cribriform plate

Prognostic factors:

1. Majority of lesions of paranasal sinuses are keratinizing sqamous cell carcinomas. These tumors are usually moderately differentiated and hence are associated with better prognosis. Non keratinizing and anaplastic carcinomas have poor prognosis.

2. Higher T stage - With increasing bulk of primary tumor, the effectiveness of irradiation takes a beating.

3. Clinically positive cervical adenopathy

4. All T4 lesions

5. Presence of distant metastasis

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Robin's clinical phases of malignant growth paranasal sinuses:

Phase I: In this phase carcinoma masquerades as sinusitis. In this phase diagnosis could be clinched by surgical procedure to secure biopsy from the suspected lesion. Drainage should also be created for the obstructed sinus cavity to drain. The presence of intractable unilateral sinusitis in a patient of susceptible age group should arouse suspicion of carcinoma

Phase II: In this phase a dull ache starts as the tumor erodes the bony walls of the nose and pns. Radiographically this phase shows a subtle asymmetry of the walls

Phase III: In this phase the nerves gets infiltrated by tumor cells leading on to hypesthesia / deep pain. This indicates advanced disease. In this phase pain may be disproportionate to the clinical findings. This phase easily gets demonstrated radiographically.

Phase IV: This phase is characterized by gross disfigurement and distant metastasis

Tumors of minor salivary glands in nose & paranasal sinuses:

Adenoid cystic carcinoma and mucoepidermoid carcinoma can occur in minor salivary glands seen in the nasal cavity and paranasal sinuses. Adenoid cystic carcinoma show solid areas of malignant cells. Vascular invasion and distant metastasis are common. Perineural invasion is also known to occur.

Sarcomas:

Osteogenic sarcomas: usually present as enlarging firm mass. This mass is clearly seen radiologically. Surgical excision may be difficult due to skull base extension.

Chondrosarcomas: Occurs rarely in nasal septum. It is difficult to differentiate from chondroma. Ring calcifications may be clearly seen in CT scans of these patients.

Soft tissue sarcomas like Rhabdomyosarcomas occur in the sinonasal area.

Patients with sarcomas are not ideal candidates for irradiation. Surgical excision should be attempted where ever possible.

Adenocarcinoma: Commonly seen in wood workers. Histologically adenocarcinomas could be classified as low grade and high grade types. High grade adenocarcinomas carry poor prognosis. These tumors commonly arise from middle turbinate. It has bone eroding properties. Nodal metastasis is also common. Surgical extripation of the tumor should be attempted where ever possible as these tumors are not radiosensitive.

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Sinonasal malignant melanoma:

These are very rare tumors of nose and paranasal sinuses. Origin from sinus mucosa is very uncommon. Survival of patients with malignant melanoma of nose and PNS is zero. Amelanotic melanomas of nasal cavity frequently present as unilateral polyps; thus it is very important to send all polypoidal material for pathological examination.

Features of malignant melanoma of Nose & PNS:

1. Most commonly these tumors arise from septum / lateral nasal wall

2. There is slight male preponderance

3. Peak age group is 5th - 8th decade

4. Frequent in orients

5. Presents as grey, blue, or black polypoidal swelling

6. Regional satellite metastasis is frequent

7. Poor prognosis

Esthesioneuroblastoma:

Synonyms: Esthesioneuroblastoma, neuroendocrine tumor

Definition: These tumors arise from olfactory epithelium in the upper nasal cavity and above the middle turbinate. It resembles anaplastic carcinoma and may be undiagnosed unless special tumor markers are used.

Cells of origin: This tumor arises from the stem cells of neural crest origin. These cells are supposed to differentiate into olfactory sensory cells. It should be regarded as one of the primitive neuroectodermal tumors. This tumor is commonly confused with small cell tumors of the nasal cavity.

These tumors appear as exophytic, polypoidal or sessile mass. The mass itself appears smooth and congested. Large tumors may present with surface ulceration. Initially these tumors are unilateral, but ultimately it extends into the opposite side of the nasal cavity and into the adjacent paranasal sinuses with continued growth.

Features of olfactory neuroblastoma:

1. Very few cases have been reported (rare)
2. Presents usually with nasal obstruction and mass
3. Almost all age groups are affected
4. Urinary VMA and Homovanillic acid cannot be usually detected
5. Shows bimodal age distribution with two peaks (around 20 and 50 years)
6. The term Neuroendocrine tumors is proposed to describe this tumor in older age group
7. Young patients show less local recurrence and more metastasis

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8. Old patients show more common local recurrence and less metastasis

These tumors are very slow growing tumors.

Clinical staging:

Group A tumors: Are confined to the nasal cavity

Group B tumors: Involves the nasal cavity and one or more of paranasal sinuses

Group C tumors: Extends beyond these limits and erodes the cribriform plate

Classification of olfactory neuroblastoma:

1. Neuroblastoma proper: This group can be subdivided into those manifesting olfactory differentiation and those without such histological evidence.

2. Neuroendocrine carcinomas

Neuroblastomas proper: Neuroblastomas without olfactory differentiation are composed of sheets of poorly demarcated groups of cells separated by fine connective tissue trabeculae. These cells are small and mitosis are rather unusual in them. The most important criteria is the presence of fibrillary material between the tumor cells. Foci of necrosis with small calcified deposits are also found. Holmer-Wright rosettes can also be seen.
These rosettes are nothing but a ring of neuroblastoma cells encircling a small space filled with neurofibrillary material.

Olfactory rosettes are the hallmark of neuroblastomas with olfactory differentiation. These rosettes are acinar spaces ringed by tall columnar cells with pseudostratified nuclei and contains small amounts of mucin and cellular debris.

Neuroendocrine carcinomas: These tumors manifest a unique feature of being admixed with glands. In some instances, the neoplastic cells are intimately related to the basal epithelium of the glands. Neurofibrillary component is not seen and the growth pattern is solid nests without rosettes.

Lymphoreticular neoplasms: Also known as Nasal T cell lymphoma. These extramedullary plasmacytoma occur commonly in the head and neck region. These lesions are usually solitary and must be considered as a tumor of well differentiated B lymphocytes. These tumors replaces rather than invade tissues. Sinonasal lymphomas are very rare tumors.

Features of Nasal T cell lymphoma:

1. Median age of involvement is 50 yrs

2. This tumor shows male preponderance

3. Carries worse prognosis than lymphoma of waldayer's ring

4. Most patients have no other evidence of lymphoma

These tumors are radiosensitive.

Figure showing growth left maxilla presenting at the gingivolabial sulcus

Management:

Management of malignant tumors of nose and paranasal sinuses is dependent on tissue diagnosis and extent of the lesion.

Extent of lesion can be assessed by clinical examination and radiological imaging.

First step is to take a complete and thorough history. Specific attention should be paid to areas adjacent to the walls of the involved sinus. The following points should be noted:

Hypesthesia / anesthesia of cheek: This indicates involvement of infraorbital nerve at the level of infraorbital foramen or in the orbit. Anesthesia of cheek is also seen in patients with tumor erosion of the anterior wall of the maxilla.

Caution * Caldwel luc surgery performed for biopsy purposes may also cause anesthesia of cheek

Numbness of upper incisors: Should be noted. If present it indicates involvement of anterior superior alveolar nerve. This is possible only if the tumor invades the floor of orbit.

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Loosening of teeth: common in tumors involving the floor of the maxillary antrum

Changing / ill fitting dentures: common in tumors involving the alveolar process of maxilla

Diplopia / ptosis: Seen when tumors invade into the orbital cavity

Trismus: is seen when the tumor invades pterygopalatine fossa

Epiphora: is seen when tumor involves the inferior meatus

Head aches commonly indicate intracranial extension

Physical examination:

Following points should be borne in mind while performing physical examination in these patients:

1. Careful examination of nasal cavity, oral cavity and nasopharynx. Mass if present should be clearly documented.

2. Medial displacement of lateral nasal wall should be looked for.

3. Inspection & palpation of maxillary teeth. This is done to look for loosening or revitalization to teeth.

4. Visualization of nasopharynx and choanae.

5. Examination of cheek and gingivolabial sulcus. Mass / fullness in the area should be looked for.

6. Involvement of infraorbital nerve should be looked for.

7. Assessment of visual acuity, ocular mobility and displacement of globe should be looked for.

8. Neck should be palpated for presence of enlarged neck nodes.

Radiologic evaluation:

There are 4 basic projections for investigating the paranasal sinuses as far as plain radiographs are concerned.

They are :

Caldwell view: This view is best for evaluation of tumors involving ethmoidal sinus and orbit.

Water's view: This view is good for evaluating maxillary sinus tumors.

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Lateral view: This view is best for evaluating tumors involving sphenoid sinus. This view also provides information about the anterior and posterior walls of frontal and maxillary sinuses.

Submental vertex view: Is best for studying tumors involving sphenoid and posterior ethmoid sinuses. This view also shows the posterior wall of maxillary sinus, pterygoid plates and the orbit.

CT scan nose and paranasal sinuses:

Both axial and cornonal sections should be studied. This imaging modality helps in accurately staging the tumor. Bone erosion if any will be clearly seen in CT scan images.

MRI scanning:

This imaging modality is complementary to CT scan imaging. MRI offers better soft tissue resolution while CT demonstrates better bone resolution. MRI is very useful in differentiating tumor mass from inflammatory lesions and retained secretions. The presence of increased water content in inflammatory lesions results in marked increase in signal on T2 weighted MRI scans. Most tumors involving the sino nasal cavities are highly cellular and demonstrate intermediate signal intensity in T2 weighted images.

Possible treatement modalitites available include:

1. Irradiation: can be used as a primary treatment modality in squamous cell carcinomas involving sino nasal cavities. This modality can also be used in conjunction with surgical extripation of the tumor mass. Irradiation should be started 6 weeks after surgery.

2. Surgery: Maxillectomy / Total maxillectomy with orbital extenteration

3. Chemotherapy

4. Combination therapy

Tissue diagnosis is important because squamous cell carcinoma and lymphomas respond to irradiation, where as adenocarcinomas and sarcomas are radioresistant.

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